Brachial artery aneurysm and thrombosis secondary to fibromuscular dysplasia

Julia Louise Jones, MBBS,[a] **Upeksha de Silva, MBBS, FRACS,**[a] and **Hwei Choo Soh, MBBS, FRCPA,**[b]
Hornsby and St Leonards, NSW, Australia

Fibromuscular dysplasia is a pathologic process causing stenosis and dilation of medium-caliber arteries of unknown etiology. It most commonly affects the renal and carotid arteries; however, it has been described in virtually all anatomic areas, including, rarely, the brachial artery. We describe a case of brachial artery aneurysm and thrombosis in a 29-year-old man secondary to fibromuscular dysplasia, treated surgically with excision, embolectomy, interposed vein graft, and anticoagulation. (J Vasc Surg Cases 2016;2:114-8.)

Fibromuscular dysplasia (FMD) is a rare arterial disorder affecting small to medium-sized arteries. It is characterized by dysplastic changes in one of the arterial layers, leading to stenosis with or without dilation. The most common form, medial fibroplasia, affects the medial layer and leads to both stenosis and dilation with a classic "string of beads" appearance on angiography. The most commonly affected arteries are the renal and carotid arteries; however, it has been documented in virtually all small- and medium-caliber arteries. The vessel involved dictates the clinical presentation of this disease; hence, renovascular hypertension and neurologic symptoms are the most common presenting complaint.[1] FMD typically affects young to middle-aged women; risk factors include family history of FMD, smoking, and hypertension. The diagnosis is confirmed on histopathologic evaluation; however, classic changes on angiography are often a substitute for tissue diagnosis. The surgical management of symptomatic or clinically significant FMD is balloon angioplasty, although open resection of the affected artery remains a treatment option in some clinical contexts.[2]

We present an unusual case of brachial artery intimal fibroplasia in a male patient with no pre-existing risk factors. The patient consented for his case to be published.

CASE REPORT

A 29-year-old man of Indian heritage with no prior medical history presented to the emergency department with 2 weeks of right arm pain and a cool right hand. He described the pain as intermittent, localizing to the hand and forearm. He had noted no paresthesia or change in sensation or motor function. He specifically had no history of hypertension, smoking, blunt trauma, or intravenous drug use. An outpatient ultrasound examination arranged by his general practitioner had shown aneurysm and thrombosis of the right brachial artery.

On review in the emergency department, the patient had mild right arm pain at rest. He was initially hypertensive, but this did not persist during admission. Physical examination showed cyanosis of the affected side, coolness of the right extremity up to the mid forearm, impalpable radial and ulnar pulses, and no palpable mass in the upper arm. His biochemical panel was unremarkable, notably with normal inflammatory markers. Bedside ultrasound showed dilation of the brachial artery and occlusion of flow 11 cm above the elbow crease. This was confirmed on computed tomography angiography, showing abrupt occlusion of contrast material in the brachial artery with reconstitution of distal flow through a small collateral vessel. Magnetic resonance angiography (Fig 1) showed a 40 × 12 × 14-mm lobulated mass lying in the course of the right brachial artery consistent with aneurysm or pseudoaneurysm and subacute thrombus. This was associated with surrounding soft tissue edema and enhancement, suggesting local inflammation.

As there was no loss of sensory or motor function, anticoagulation with intravenous heparin was commenced overnight with operative management undertaken the following day. The 35-mm aneurysmal segment of the brachial artery was resected (Fig 2) and embolectomy performed. Concurrently, a segment of the right saphenous vein was harvested and used as a reversed venous interposition graft. After completion of the graft anastomosis, intraoperative ultrasound showed clot and obstruction of the distal radial and ulnar arteries. An arteriotomy of the brachial artery, just above the bifurcation, was performed. An embolectomy of the distal brachial, radial, and ulnar arteries was successful. Subsequently, there was improvement in pallor, warmth, and capillary refill of the distal extremity, although the radial pulse remained difficult to palpate. Anticoagulation initially continued postoperatively; however, this was stopped because of hematoma formation requiring a return to the operating room for clot evacuation.

Histopathologic analysis of the resected artery found evidence of subacute aneurysm rupture with organizing hematoma.

From the Department of Surgery, Hornsby Kuring-Gai Hospital, Hornsby[a]; and the Department of Anatomical Pathology, Royal North Shore Hospital, St Leonards.[b]

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Correspondence: Julia Louise Jones, MBBS, Department of Surgery, Hornsby Kuring-Gai Hospital, Palmerston Road, Hornsby, NSW 2077, Australia (e-mail: jjon9461@uni.sydney.edu.au).

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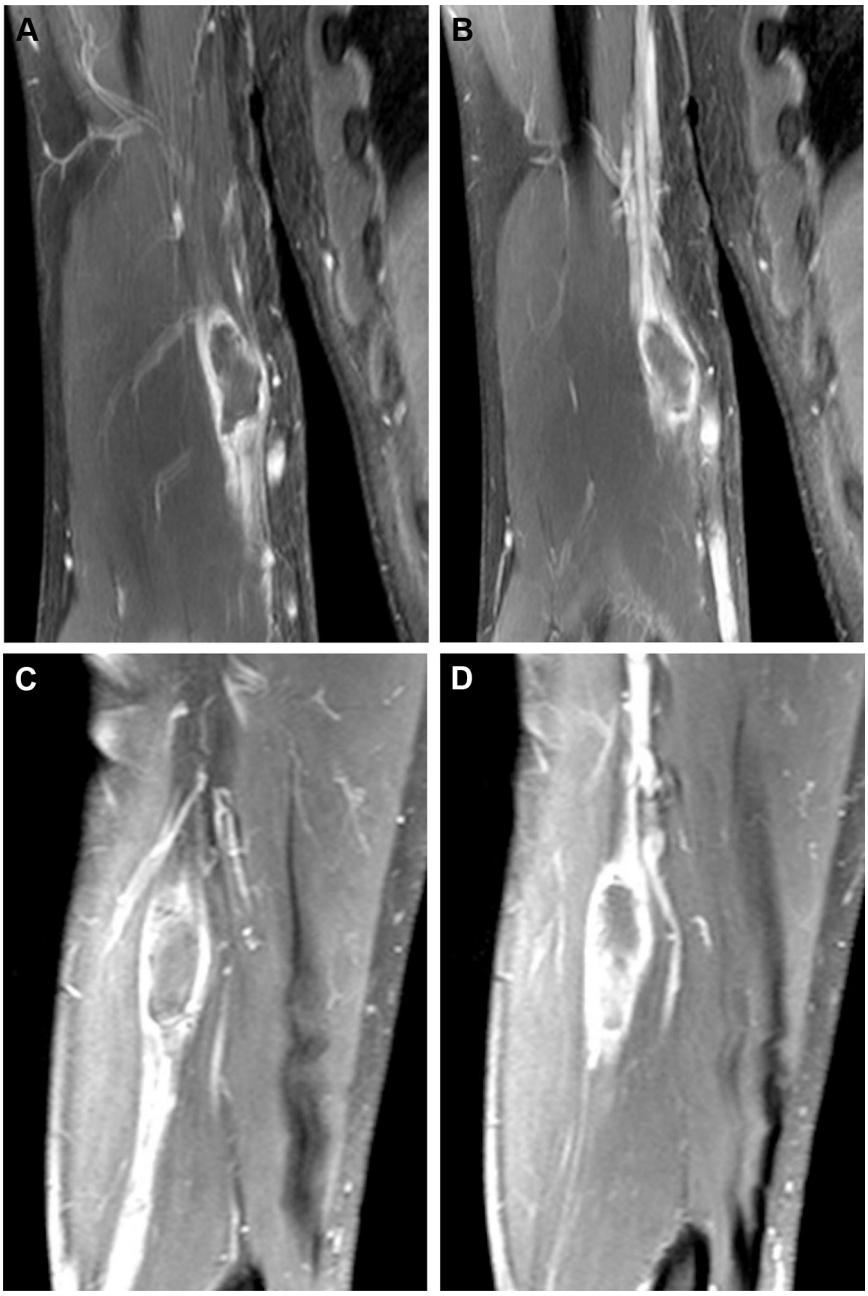

Fig 1. Magnetic resonance angiography with gadolinium contrast enhancement of the right upper arm showing an aneurysm with subacute thrombus obstructing flow. **A** and **B,** Coronal view. **C** and **D,** Saggital view.

The arterial wall displayed thickening of the intima, fragmentation of the internal elastic lamina, and a thinned and disorganized media alternating with thickened nodular areas, intervening fibrosis, and loss of smooth muscle, creating an undulating effect on the vessel wall (Fig 3). There was a notable absence of inflammatory or fatty infiltrates. These findings were reported as consistent with a dysplastic process such as FMD.

He was discharged 7 days after admission with palpable brachial and ulnar pulses and radial pulse detectable on Doppler ultrasound. He was followed up as an outpatient at 6 weeks. He had no ongoing arm pain. Doppler ultrasound showed that the graft was widely patent; the radial artery was patent, but the ulnar artery was narrowed and occluded at the wrist. Photoplethysmography was performed on the right-side digits and was normal. A review of the initial computed tomography angiogram showed no other arterial abnormalities, suggesting no concurrent FMD of the contralateral brachial, renal, or extracranial arteries. The intracranial arteries are yet to be screened for FMD.

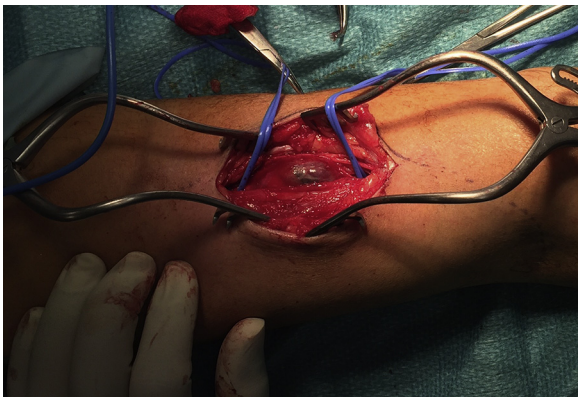

Fig 2. Intraoperative image of skeletonized aneurysm before resection.

DISCUSSION

Whereas FMD is uncommon, FMD of the brachial artery is very rare, with only 29 cases of unilateral or bilateral brachial artery involvement reported in the literature since 1984 (Table). These cases were largely diagnosed after investigations for upper limb ischemia or were incidental findings on imaging or angiography. Although it is uncommon, FMD remains an important differential diagnosis in the evaluation of arterial insufficiency and stenosis.

FMD can typically be distinguished from atherosclerotic disease by the lesion's location and lack of vascular risk factors. FMD affects the mid to distal segments as opposed to the proximal narrowing seen in atherosclerotic lesions. It can be distinguished from inflammatory vasculitides by the absence of elevated serum inflammatory markers and inflammatory infiltrates on histopathologic evaluation. However, difficulty arises in the approximately 40% of vasculitides in which serum inflammatory markers are not elevated, and it can be difficult to distinguish these entities with radiologic methods alone.[1] Without tissue diagnosis, this case would have been difficult to definitively diagnose because of the nonclassic findings of aneurysm and occlusion on initial imaging.

Furthermore, the case described is unusual in that although it presented with symptoms of upper limb ischemia, the initial culprit appeared to be an aneurysm leading to the patient's symptoms. True brachial artery aneurysms are also extremely rare; they are more commonly pseudoaneurysms secondary to trauma of the artery, that is, secondary to blunt trauma, iatrogenic instrumentation,

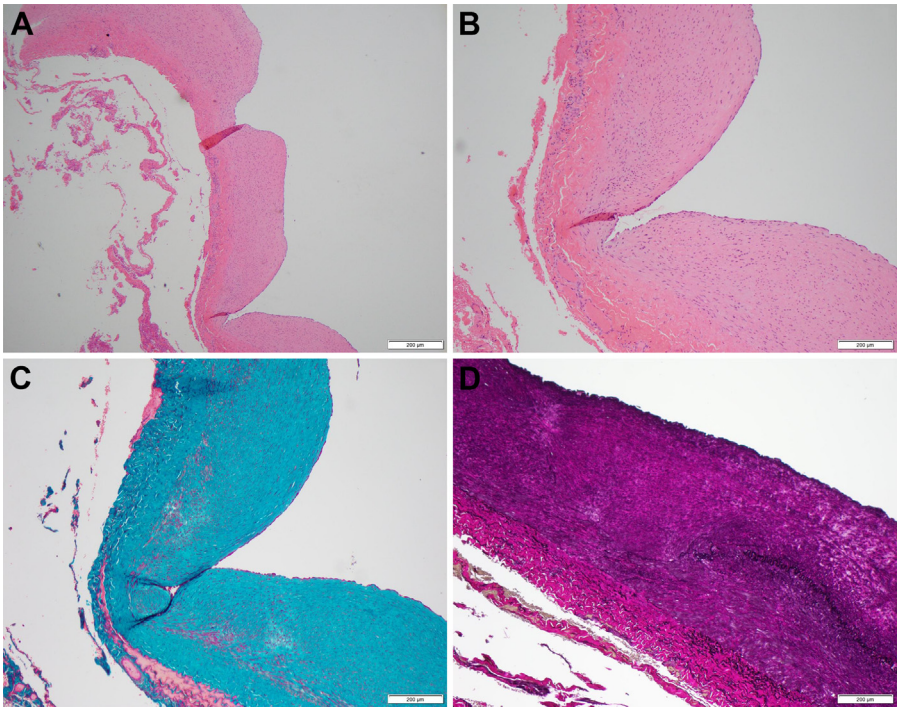

Fig 3. Histologic images, intimal surface on the upper right side. **A,** Low-power view of wall of the artery showing an undulating appearance. There is thickened intima with thinning and disorganization of the muscularis (hematoxylin and eosin stain, magnification ×50). **B,** Higher power view of wall of the artery showing **(A)** in greater detail (hematoxylin and eosin stain, magnification ×100). **C,** Wall of the artery showing thinning and disorganization of the muscularis (cells with *red* cytoplasm; Masson trichrome stain, magnification ×100). **D,** Wall of the artery showing disruption of the internal elastic lamina (*black*; Verhoeff-van Gieson stain, magnification ×100).

Table. Summary of fibromuscular dysplasia (FMD) cases affecting the brachial artery

First author, year	*Age, years, and sex*	*Presentation*	*Side affected*	*Treatment*
Olson, 1984	42 F	LUL intermittent paresthesia	Left	Rxn, L brachial RSVG
Iwai, 1985	25 F	RUL first and second digit cold and pulseless	Right	Stellate ganglion block
Iwai, 1985	64 F	Incidental finding in patient with claudication	Unspecified	No treatment
Esfahani, 1989	22 M	RUL tissue loss	Right	Not described
Esfahani, 1989	37 M	Symptomatic	Unspecified	Not described
Cheu, 1991	74 F	RUL second digit pain and tissue loss	Right	Rxn, R brachial RSVG
Lin, 1992	74 F	RUL third digit pain and tissue loss	Right	Rxn, R brachial RSVG
Reilly, 1993	77 F	RUL cyanosis and decreased pulses	Bilateral	Rxn, R brachial RSVG and no treatment on L
Shipolini, 1993	63 F	RUL pain and exercise-induced paresthesia and weakness, aneurysm	Right, recurrent	Rxn, R brachial RSVG
Dorman, 1994	64 F	RUL second digit pain and discoloration	Bilateral	Rxn, R brachial RSVG and no treatment on L
Yoshida, 1994	45 M	LUL pain and cyanosis of fingers	Left	L PTA
Ciocca, 1995	63 F	LUL pain, coldness, and paresthesia	Left	Infusion with urokinase, L PTA
Haueisen, 1998	51 F	LUL third digit pain, coldness, and reddening	Left	Rxn, L brachial RSVG
Haueisen, 1999	54 F	LUL pain, coldness, and paresthesia	Left	Rxn, L brachial RSVG
Suzuki, 1999	65 F	Incidental finding during heart catheterization	Bilateral	Not described
Suzuki, 1999	89 F	Bilateral coldness of fingers during winter	Bilateral	Not described
Vuong, 1999	59 F	Workup of left brachial artery pseudoaneurysm	Left	Rxn, L brachial RSVG
Cutts, 2000	62 F	BUL paresthesias, pain, and bruits	Bilateral	Rxn, R and L brachial RSVG
Nozaki, 2003	8 months M	RUL delayed growth plus constellation of neurologic signs	Right	Not described
Kolurri, 2004	61 F	RUL tissue loss, LUL incidentally on heart catheterization	Bilateral	PTA
Shin, 2007	61 F	RUL pain, paresthesia, weakness, and radial embolus; LUL incidental	Bilateral	Rxn, R brachial RSVG and thrombolysis, and no treatment on L
Yoshimuta, 2008	56 F	Incidental finding during heart catheterization	Bilateral	Not described
Ministro, 2008	69 F	RUL second digit pain and tissue loss	Right	Rxn, R brachial RSVG
Margoles, 2009	83 F	LUL arteriovenous graft dysfunction	Left	PTA
Rice, 2010	76 F	RUL second digit pain, paresthesia, and tissue loss	Right	Rxn, R brachial RSVG
Lewis, 2011	65 F	BUL exertional ischemia	Bilateral	PTA
De Waele, 2012	46 M	RUL pain and pseudoaneurysm workup	Right	Rxn, R brachial RSVG
Kar, 2013	58 F	Incidental finding in stroke and aortic dissection workup	Right	No treatment of brachial component
Liang, 2014	—	Incidental finding on CTA after spontaneous coronary artery dissection	Unspecified	Not described

BUL, Bilateral upper limbs; *CTA,* computed tomography angiography; *LUL,* left upper limb; *PTA,* percutaneous transluminal angioplasty; *RSVG,* reversed saphenous vein graft; *RUL,* right upper limb; *Rxn,* resection.

or intravenous drug abuse, none of which were present in this patient's history. Macroaneurysms and dissections are acknowledged as complications of FMD.[2] Two previous cases of FMD were discovered after investigation and treatment of brachial artery pseudoaneurysm.[3,4] There is only one previously reported case of true brachial artery aneurysm secondary to FMD.[5]

The histopathologic findings in this case are consistent with the less common form of FMD, intimal fibroplasia, found in <10%. On histologic evaluation, this is characterized by the absence of lipids or inflammatory cells; collagen deposition in the intima, which may be circumferential or eccentric; and fragmentation or duplication of the internal elastic lamina.[6] On angiography, rather than the string of beads appearance typically seen, the affected artery may have a concentric focal band or long smooth narrowing.[1]

Importantly, because FMD affects multiple vascular beds in 28% of cases,[2] screening of the most commonly affected sites, the renal and cervicocerebral arteries, should be undertaken.

CONCLUSIONS

This case reminds us that although FMD is a rare disease, rarely affecting the upper limb, it remains an important differential diagnosis in arterial insufficiency.

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